Primary myxoma of the parafalcine meninges

Case report

CIARAN J. POWERS, M.D., PH.D., CATHERINE C. PIZZI, M.S., THOMAS J. CUMMINGS, M.D., AND ALLAN H. FRIEDMAN, M.D.

Division of Neurosurgery, Department of Surgery, and Department of Pathology, Duke University Medical Center, Durham, North Carolina

The authors report on an unusual case of a primary intracranial myxoma in a 39-year-old woman. The patient presented with headache and generalized seizure. Magnetic resonance imaging revealed a large right frontal tumor resembling a parasagittal meningioma. A gross-total resection was performed, and histological review confirmed the lesion as a myxoma. Results of additional workup revealed the absence of a primary myxoma elsewhere. This case represents the third published report of a primary intracranial myxoma and the second report of a supratentorial myxoma.

**Key Words** • myxoma • central nervous system • meningioma

**Abbreviation used in this paper:** MR = magnetic resonance.
was discharged home on postoperative Day 2. She was seen in the clinic 3 weeks after surgery, and her neurological condition remained normal. A postoperative MR image confirmed gross-total resection of the tumor. After a final review of the pathological study revealed myxoma, the patient underwent additional workup including echocardiography, which revealed no other primary source for the intracranial tumor.

**Discussion**

Primary intracranial myxoma is a very rare entity, with only three case reports in the literature. In contrast, seeding of the brain by tumor emboli from a primary cardiac myxoma is a well-described phenomenon. These so-called metastatic myxomas occur with a reported incidence of 27 to 50% in patients with atrial myxoma. Interleukin-6 may play a causative role in the metastasis of cardiac myxomas. Myxoid emboli from a cardiac source have also been associated with aneurysms of the intracranial circulation.

The first report of a primary intracranial myxoma was made by Nagatani, et al., in 1987. These authors presented the case of a 38-year-old man who initially suffered rapid right visual loss. Imaging studies revealed an enhancing mass based on the right side of the pituitary fossa with suprasellar extension. An extensive endocrine workup was normal. The tumor was resected via a transsphenoidal approach, and histological examination revealed myxoma. Echocardiography demonstrated no evidence of primary cardiac myxoma, and the patient had no sign of recurrence 4 years after tumor resection. These authors proposed a tumor origin from primitive mesenchymal cells in the synchondrosis of the sphenoid bone.

In 1990 Klein, et al., presented a case of primary intracranial myxoma of the posterior fossa. They described a 32-year-old woman who had presented with a 3-week history of headaches. Computerized tomography scanning revealed an inhomogeneously enhancing 4-cm mass in the left posterior fossa, and at surgery the tumor was found to be attached to the dura mater. Histological examination revealed myxoma with no meningeal, neural, or glial components. Echocardiography showed no evidence of a primary cardiac myxoma. Immediately postoperatively the patient was neurologically normal; no long-term follow-up information was reported.

Graham, et al., presented a case of primary supratentorial myxoma in 1999. These authors described an adolescent female who had presented with a 3-day history of headache and intermittent left-sided focal motor seizures. Computerized tomography scanning and MR imaging revealed a 3 × 5-cm inhomogeneously enhancing extraaxial mass in the right frontal region, consistent with a convexity meningioma. At surgery a dural-based mass was resected, and subsequent histological examination revealed the tumor to be a myxoma. The authors examined the tumor specimen by using electron microscopy and found slender bipolar mesenchymal cells without features of meningothelial cells, con-
sistent with myxoma rather than meningioma with myxoid degeneration. Echocardiography studies were negative. At the 5-year follow-up examination the patient remained neurologically normal. The authors proposed that the tumor had arisen from the primitive mesenchymal elements of the convex dura.

Graham, et al., generated differential diagnoses for myxoid intracranial tumors including myxomatous meningioma, epithelioid hemangioendothelioma, sarcoma with myxoid degeneration, and metastatic myxoma. Additional intracranial myxoid tumors include fibrous histiocytoma and intracranial neurothekeoma. Differentiation among these tumors is very important insofar as additional workup is required to find a primary cardiac tumor if the lesion is a true myxoma. Immunohistochemistry is the most useful tool for differentiating these lesions. Meningioma with myxoid components will stain positive for epithelial membrane antigen, sarcoma shows positive staining for smooth muscle actin and desmin, and neurothekeoma will stain positive for $\text{S100}$ protein. True myxoma will not stain with any of these markers but will stain positively for vimentin, which is consistent with its probable mesenchymal origin. Because of the possibility of sampling errors, it is important for the entire tumor to be examined for any elements consistent with meningioma, sarcoma, or other tumor prior to the diagnosis of myxoma. Once the diagnosis of myxoma is made, echocardiography is vital in ruling out a cardiac myxoma as the primary tumor. Histological examination may be suggestive of a primary intracranial myxoma if the lesion arises from the meninges rather than from small blood vessels supplying the brain parenchyma, as would be expected with a metastatic tumor.

As with the patient in the report by Graham and colleagues, our patient presented with headaches and seizures. In addition, initial imaging studies were consistent with a supratentorial meningioma having dural attachment. The tumor was completely resected, and the patient has continued to do well postoperatively.

**Conclusions**

An unusual case of a primary supratentorial myxoma presenting as a parafalcine meningioma is reported. The patient underwent gross-total resection, which was confirmed on postoperative imaging. No extracranial source of the tumor was found. Although cases are few, the prognosis for fully resected primary intracranial myxoma is thought to be excellent. Echocardiography is an important diagnostic test to rule out a primary cardiac tumor. The origin of these tumors remains unknown, although it is likely that they arise from primitive mesenchymal cells.

**References**


---

**Fig. 2.** A: Representative photomicrograph of myxoid tumor section demonstrating entrapped stellate and elongated cells with whispy eosinophilic processes and elongated bubbly nuclei lacking mitotic activity. In some areas the vascularity is increased. H & E, original magnification $\times 25$. B: Representative photomicrograph of myxoid tumor section revealing brilliant blue staining, which indicates an abundance of mucin in the tumor and is consistent with the diagnosis of myxoma. Alcian blue, original magnification $\times 50$.  

---

Primary intracranial myxoma


Manuscript received October 8, 2005.
Accepted in final form December 1, 2005.
Address reprint requests to: Allan H. Friedman, M.D., Division of Neurosurgery, Duke University Medical Center, Box 3807, Durham, North Carolina 27710. email: fried010@mc.duke.edu.